ANGIOMATOID MALIGNANT FIBROUS HISTIOCYTOMA

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A number of cases of tumors of fibrous and histiocytic nature with malignant potential are reported(1). These lesions are generally in the 5-7 decades, but a few instances in children have been recorded(2). A variant of malignant fibrous histiocytoma had been described by Enzinger(3), an angiomatoid malignant fibrous histiocytoma (AMFH) which shows differences from usual forms of the neoplasm. We report a child with such a rare type of tumor.

Case Report

An 8-year-old boy was admitted in S.S.K.M. Hospital, Calcutta with a swelling over the left side of the neck, left sided facial palsy, neck stiffness, low grade fever and extreme emaciation for 3 months. The child had been suffering from low grade irregular fever for a variable period, loss of weight and neck stiffness six months prior to hospitalization and was treated in a nursing home with antitubercular drugs. Though there was some alleviation of his constitutional symptoms but no reduction of neck stiffness was noted inspite of continuation of these drugs. About 3½ months ago, the patient had a minor fall following which he developed a small painless hard nodular swelling over the left upper posterior part of the neck which had rapidly enlarged to its present size and facial palsy on the affected side.

On examination the child was grossly emaciated, pale, weighed 10 kg and had a midarm circumference of 9.5 cm. A big nodular lump 4 cm × 4.5 cm was palpable over the upper part of the left side of the neck. The lump was hard with cystic feel at the centre, ovoid in shape with diffuse margin, non-tender and fixed to the deeper structures. Skin over the swelling was fixed. There was also lower motor neurone type of facial palsy on the left side.

Investigations showed a hemoglobin level of 7.5 g/dl, total leucocyte count 9,500/cu mm, differential count of 37% polymorphonuclear cells 53%, lymphocytes 10%, eosinophils and adequate number of platelets. Mantoux test was negative and no accelerated reaction was noted after BCG vaccination. Roentgenography of skull, mastoid, cervical spines and chest revealed no abnormality. CT scan of cranium showed a large soft tissue mass at the left mastoid region with normal brain parenchyma. A fine needle aspiration cytology of the tumor revealed cluster of cells with intercellular collagen. The cells were of two types–one with elongated nuclei and scanty cytoplasm and other with eccentric ovoid nuclei and abundant cytoplasm. Multinucleated giant cells and bizarre cells were present which was suggestive of malignant fibrous histiocytoma. Biopsy from the tumor showed nodular masses of malignant fibroblasts and histiocyte-like cells in relation to blood filled

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spaces. A lymphocytic and plasma cell infiltration were seen around the nodules (*Fig.*). These histological findings were consistent with the diagnosis of malignant fibrous histiocytoma (antgiomatoid type).

The patient was treated with intravenous administration of vincristine, cyclophosphamide, doxorubicin and actinomycin D. Radiotherapy could not be given due to critical condition of the patient. The boy succumbed 8 weeks after hospitalization.

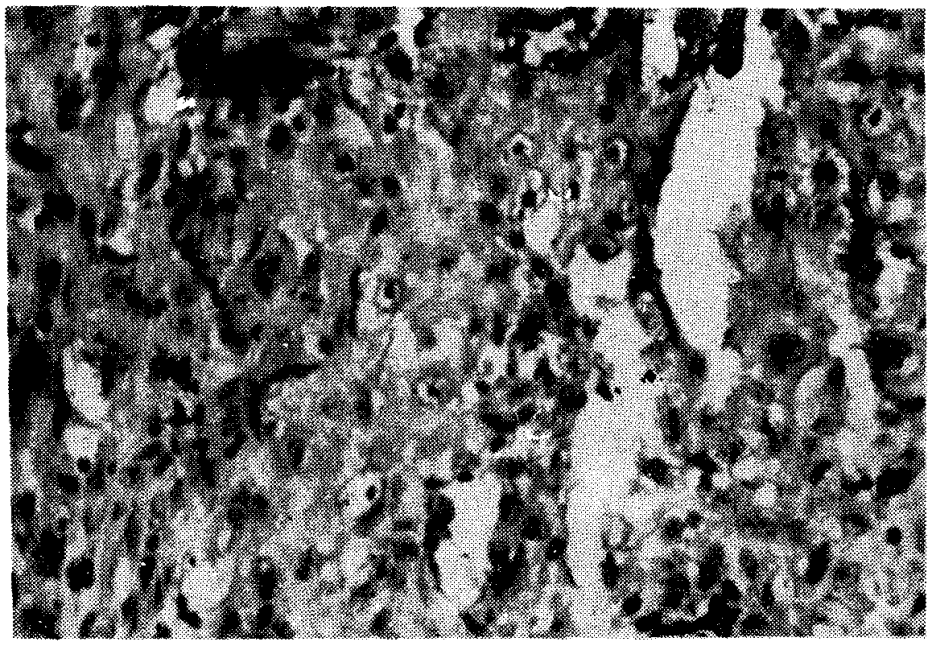

Fig. Microphotograph showing dual cellular composition of the tumor. Malignant spindle shaped fibroblasts are seen admixed with large polygonal histiocyte like cells. There are also vascular clefts containing RBC (H&E x 320)

Discussion

AMFH is a well defined clinicopathological entity which affects mainly children and young adults(3-4). They are found mainly in the extremities and appear as painless subcutaneous nodules varying between 2 to 10 cm in diameter(3). The tumors are highly vascular and show infiltration of the surrounding tissue. Systemic effects of the tumor include anemia, weight loss and fever. Though AMFH is less aggressive than other varieties of malignant fibrous histiocytoma, 12 of Enzinger's 24 patients had recurrences and 5 had distant metastasis(3). A larger lesion of similar type, reported by Santa-cruz and Kyriakos(5), was regarded as a subtype of fibro-histiocytic tumors. Sun(6) suggested the tumor as vascular in origin perhaps related to a hemangioma. Lower motor neurone type of facial palsy was most probably due to pressure effect of the lump at the nerve exit. Unfortunately, post-mortem confirmation of the clinical findings could not be ascertained in this case.

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PULMONARY ALVEOLAR PHOSPHOLIPOPROTEINOSIS

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Primary alveolar proteinosis (PAP) now phospholipoproteinosis is a syndrome of unknown etiology described by Rosen and *et al.* in 1958(1). PAP results from abnormal accumulation of surfactant phospholipids and proteins in the alveolar spaces. The disorder has been described in all the age groups from six months to 72-year-old patients, but the preponderance of those affected is between 20 and 50 yrs(2). However, the number of cases occurring in infants and children among the total cases described is small(3) and is now being recognized as a possible cause of neonatal respiratory distress(4).

Case Report

A 14-year-old school student was admitted with the complaints of reduced appetite, failure to gain weight and easy fatigability since the age of 2 years. The patient also complained of progressively increasing exertional dyspnea since the past 2 years and dry cough for the past 6 months. He was treated with antituberculous treatment for four months by a private practitioner which was omitted due to lack of clinical and radiological response. The child was a full term normal hospital delivery, with birth weight of 2.5 kg. He was fully immunized. There was no significant past and family history.

The child was undernourished, asthenic with height of 142 cm and weight of 27 kg (expected weight per ICMR chart is 48 kg). There was no clubbing or significant lymphadenopathy. Respiratory system examination revealed occasional end inspiratory rales with bilateral scattered rhonchi. Other systems' examination was essentially normal.

His complete hemogram, renal and liver function tests were normal. Serum LDH level was normal. *X*- ray chest showed bilateral fluffy alveolar shadows distributed around both the hilar and perihilar region extending into all the zones. The shadows were static when compared with the previous roent-

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